

Case Report

DOI: <http://dx.doi.org/10.18203/2349-2902.ijssurgery202867>

Presacral schwannoma: a bizarre presentation in the Pandora's box of abdomen and pelvis

Harsha S. Poojary^{1*}, Vishwanath Sherigar², Abhinav Balaji³

¹Department of General Surgery, A.J. Institute of Medical Sciences and Research Centre, Mangalore, Karnataka, India

²Department of Oncosurgery, ³Department of Urology, A.J. Hospital and Research Centre, Mangalore, Karnataka, India

Received: 07 May 2020

Accepted: 11 June 2020

***Correspondence:**

Dr. Harsha S. Poojary,

E-mail: poojaryharsha92@gmail.com

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ABSTRACT

Schwannomas are benign tumors arising from the Schwann cells of nerve fibers. They are extremely rare in the pelvis accounting for only 1-3% of all schwannomas. These tumors are nonaggressive, slow growing, solitary neoplasms with an extremely low possibility of malignant transformation or recurrence after excision. We present a case of a 19 years old male with complaints of radiating pain from lower back to the left thigh associated with altered bowel and bladder habits. Following a detailed work up he underwent laparotomy and mass excision. Histopathological report revealed presacral schwannoma. Post-surgery patient improved symptomatically and is on regular follow up. There are a smaller number of cases of presacral schwannoma reported in the literature. Due to its outlandish striking presentation, diagnosis is very challenging. Although presacral schwannoma is rare, it should be considered as a differential diagnosis in the back of the mind of a surgeon while dealing a case of pelvic mass. In symptomatic and asymptomatic cases, surgical excision is the mainstay of treatment of these tumors.

Keywords: Presacral schwannoma, Ancient schwannoma, Antoni A, Antoni B, SMARCB1, LZTR1 mutation

INTRODUCTION

Schwannoma also known as neurilemmoma are benign, well encapsulated tumors arising from Schwann cells. Schwannomas are the commonest benign tumours representing approximately 8% of all soft tissue neoplasms. They commonly arise from head and neck, mediastinum and extremities. Pelvis is an extremely rare site of occurrence for schwannoma. Approximately 1-3% of the schwannoma arise from the pelvis and only a handful gets reported.¹

The patients are usually asymptomatic unless there is a large mass arising from the pelvis causing symptoms like that of constipation or bladder disturbances due to extrinsic compression effect or complaints of low back ache as a result of nerve root compression (sciatica).

Other differential diagnosis of a pelvic mass to be considered are vestigial congenital tumors (teratoma), vestigial cysts (dermoid, epidermoid) or non-vestigial formations (meningocele, hamartoma, chordoma), bone tumors (chondrosarcoma, Ewing's tumor, myeloma, osteosarcoma) nerve tumors (ependymoma, neurofibroma, ganglioneuroma) and soft tissue sarcoma (including malignant schwannoma).² In the following case report we discuss about a 19 years old male with a pelvic mass.

CASE REPORT

A 19 years old male presented to the department of oncosurgery with complaints of radiating pain from lower back to the left thigh, difficulty in micturition and defecation since, 2 months. On digital rectal examination, a large, immobile mass was noted. Routine blood workup

was within normal limits. CT and MRI pelvis showed a well encapsulated mass with no local vascular invasion in the pelvis into the sciatic foramen on the left side (Figure 1 and 2).



Figure 1: MRI showing a well encapsulated mass seen in the pelvis extending via the left sciatic foramen.



Figure 2: MRI pelvis (cross-section) showing the pelvic mass.



Figure 3: Lesion noted in the pelvis intra-operatively.

Patient underwent laparotomy and pelvic mass excision. Intraoperative findings noted approximately 20x20cm well encapsulated solid cystic mass arising from the presacral plexus on the left side abutting the left external and internal iliac artery and vein and its branches (Figure 3-5). Histopathology of the specimen showed spindle Schwann cell fascicles forming cellular verocay bodies (Antoni A pattern) and loosely arranged myxoid area (Antoni B pattern) (Figure 6 and 7).

Post-surgery patient condition improved. Patient was on regular follow up and had endorsed drastic reduction in the radiating pain from the lower back and no altered bowel or bladder habits.

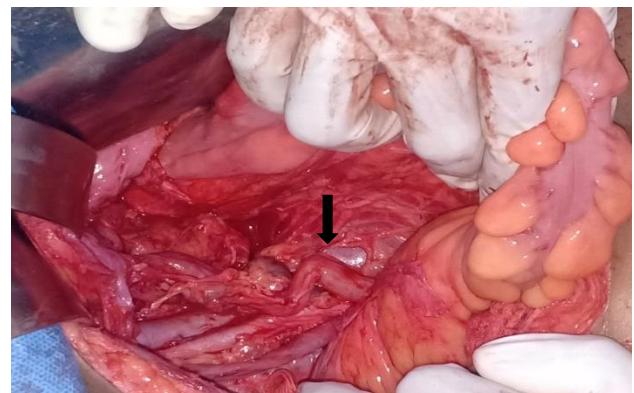


Figure 4: Tumor bed showing the left sided external and internal iliac vessels.



Figure 5: Gross specimen showing a well encapsulated mass.

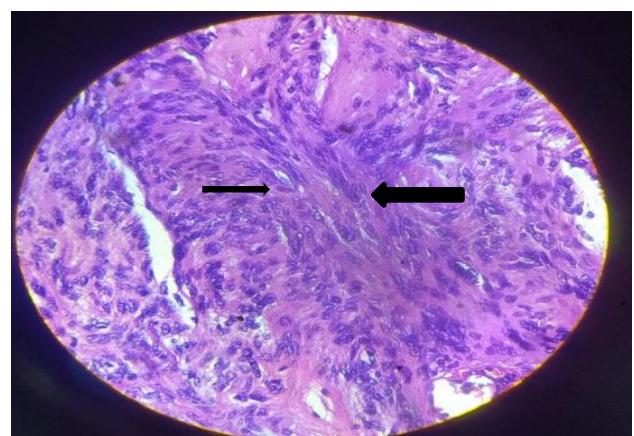


Figure 6: Histopathological examination of the specimen - small arrow pointing towards verocay bodies, big arrow pointing at the cigar shaped nuclei.

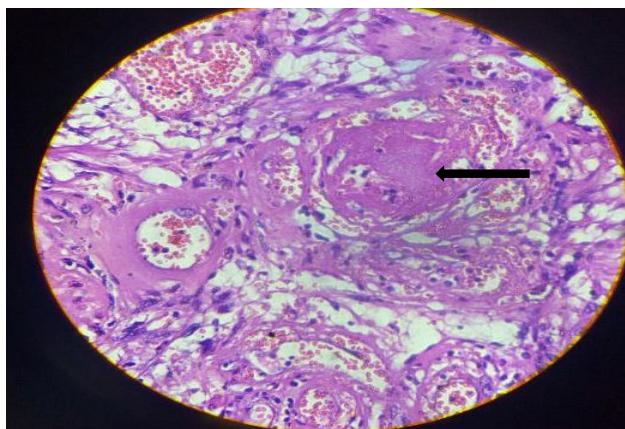


Figure 7: Histological specimen showing areas of hyalinization.

DISCUSSION

Schwannomas are benign tumors arising from the nerve sheath. Predominantly seen in men of 30-50 years of age. Schwannoma arising from the pelvis account for only 1-3 % of the tumor. Molecular studies show that certain tumor suppressor genes like SMARCB1 and LZTR1 are responsible for development of schwannomas.² Three types of pelvic schwannoma have been described: retroperitoneal or presacral schwannoma, intraosseous schwannoma or spinal schwannoma.³ Klimo and colleagues divided pelvic schwannoma into three groups: purely sacral/intracanalicular (type I), purely presacral (type III), or mixed intracanalicular/presacral (type II).⁴ Due to extensive number of differential diagnosis, many a times misdiagnosis of pelvic schwannomas as psoas abscesses have also been noted.⁵

Pre-operative diagnosis is difficult since it's a rare entity with nonspecific imaging features on CT and MRI. The imaging study in this case showed a well encapsulated solid mass in the Presacral region. Hence, we had a working diagnosis of schwannoma, Paraganglioma or a Neurofibroma. To confirm the diagnosis, histopathological examination was done. Histologically, typical schwannomas are composed of intermixed Antoni A (highly ordered cellular component) and Antoni B (myxoid hypocellular component).⁶

Complete surgical excision of the mass is the mainstay of treatment. Due to the benign nature of the lesion no adjuvant treatment is required. Local recurrence was noted wherever incomplete resection was done. Prognosis is always good in case of benign schwannoma.

CONCLUSION

Presacral schwannomas are a rare entity with an extensive differential diagnosis. Usually the diagnosis is delayed due to the late onset of symptoms. Once a presacral

mass is identified, whether benign or malignant; an aggressive approach is required to ensure en-bloc resection while maintaining the integrity of the pelvic structures. This would prevent the recurrence and improve the survival.

ACKNOWLEDGEMENTS

I take this opportunity to thank my co-author Dr. Vishwanath, consultant oncosurgeon, Department of Oncosurgery, A.J Hospital and Research Centre, Mangalore for his guidance, Dr. Sunil Shetty, consultant neurosurgeon, Department of Neurosurgery, A.J Hospital and Research Centre, Mangalore, Dr. Abhinav Balaji, senior resident, Department of Urology, A.J Hospital and Research Centre, Mangalore for their valuable inputs and Dr. Muktha Pai, Professor and Head of the department of Pathology, A.J. Institute of Medical Sciences and Research Centre for providing us with a detailed histopathological report along with the images. Lastly, I thank my patient and all the staff members in the department who have helped me during the formulation of my case report.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Poojary HS, Sherigar V, Balaji A. Presacral schwannoma: a bizarre presentation in the Pandora's box of abdomen and pelvis. *Int Surg J* 2020;7:2439-41.